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Oligodendroglioma Arising in a Sacrococcygeal Immature Teratoma

Tumors of neuroepithelial origin are extremely rare in teratoma and tend to be derived from glial or primitive neuroectodermal cells. We describe a case of 2-month-old baby girl with an oligodendroglioma arising in an immature teratoma of the sacrococcygeal region. Histologically, the tumor was identical in appearance to low grade oligodendroglioma within the adult brain. Because immature teratoma was grade II, the patient received adjuvant chemotherapy. The patient died of progression of the intra-abdominal tumor 6 months after surgical excision. The authors believe this to be the first presentation in the world literature.

Key Words : *Sacrococcygeal Region; Teratoma; Oligodendroglioma*

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INTRODUCTION

Tumors of neuroepithelial origin may rarely arise within teratoma. Only a case of teratoma with myxopapillary ependymoma has been described (1). Oligodendroglioma has not been reported within the teratoma. We report a case of an immature teratoma with oligodendroglioma, which underwent diagnosis at 2 months of age.

CASE REPORT

A 2-month-old baby girl presented with a large, non-tender swelling of the right buttock. The baby was born by a cesarean-section delivery at 40 weeks' gestation, birth weight 3.60 kg due to cephalopelvic disproportion. She had a visible mass at birth over the sacrococcygeal area, which had gradually increased in size. The pelvic computed tomography showed a huge, inhomogenous mass displacing the urinary bladder anteriosuperiorly in the presacral space. Serum level of alpha-fetoprotein (AFP) was elevated for her age (1,004.2 ng/mL). The tumor was completely removed by surgery. On gross examination, the specimen was a well-circumscribed, non-encapsulated, solid mass measuring $10.0 \times 10.0 \times 8.0$ cm and weighing 285 g, which was attached to the surrounding skeletal muscle. Sectioning revealed a relatively well delineated, soft, and gelatinous mass, 3.0×3.0 cm, on teratomatous background (Fig. 1). On histopathological examination, the teratomatous component showed keratinized stratified squamous epithelium, skin adnexa, adipose tissue, connective tissue, hyaline cartilage, glandular tissue, pseudostratifed ciliated respiratory epithelium, and immature neural tissue. Primitive neuroepithelial elements mimicking the embryonic neural tube were found in two low-power fields in the one slide. Focal ganglionic differentiated areas were found (Fig. 2). The gelatinous mass showed slightly nodular arrangement. Scattered calcospherites and plexiform, "chicken wire"-like network of thin walled blood vessels were found. The tumor cells showed uniformity of nuclear size and shape, and were admixed with angulated small vessels. The uniform, round nuclei, containing finely granular chromatin were surrounded by a clear halo of cytoplasm (Fig. 3). No mitotic activity, endothelial hyperplasia, or necrosis was identified. These findings were consistent with low grade oligodendroglioma. The tumor was diagnosed as low grade oligodendroglioma arising in an immature teratoma of the sacrococcygeal region, grade II. The patient received adjuvant chemotherapy with vincristine, VP16, and cisplatin. At systematic follow-up, the levels of AFP remained elevated for age.

Three months later, she was readmitted due to distended urinary bladder and urinary tract infection. Pelvic ultrasonogram revealed a round, heterogenous, and hyperechogenic mass, measuring 3.5×3.2 cm, in the posterior wall of the uterus, located anterior to the site of the initial tumor. Abdominal magnetic resonance imaging revealed multiple hepatic nodules. Serum AFP level elevated at 1,297.2 ng/mL. After follow-up chemotherapy, the patient developed fever and leukocytopenia. She died of sepsis and progression of the intra-abdominal tumor 6 months after the surgery. No autopsy was performed.

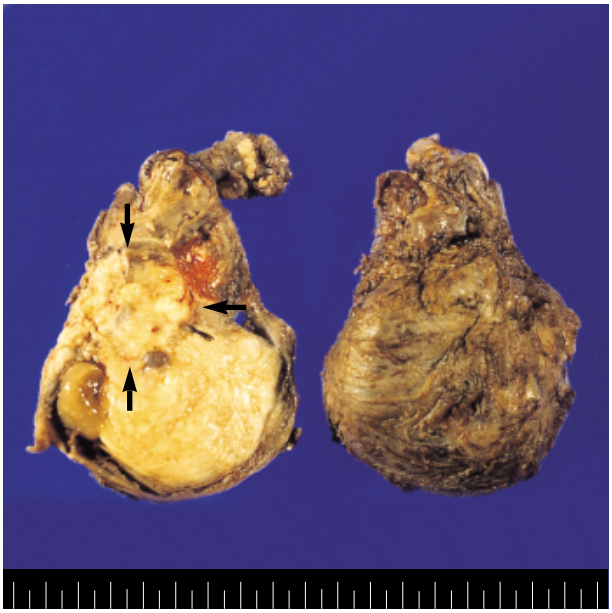

Fig. 1. The teratoma mass is well circumscribed, solid mass with a variegated surface. The cut surface reveals a relatively well delineated, soft, and gelatinous mass within the teratoma (arrows).

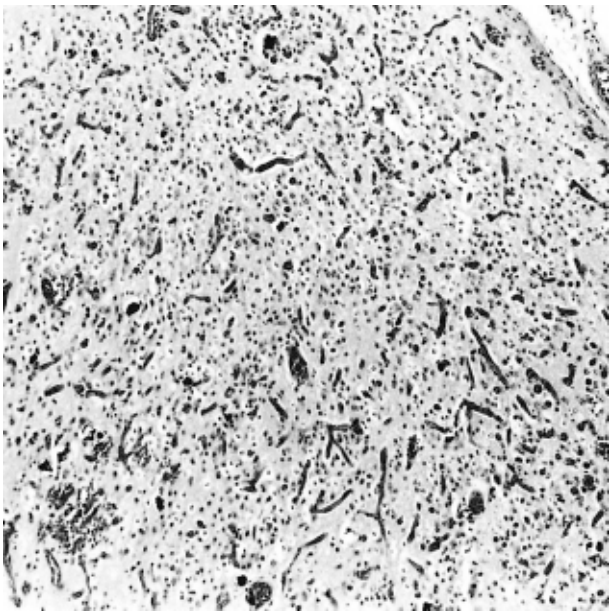

Fig. 3. The tumor cells have uniform, round nuclei and clear perinuclear halos. Scattered calcospherites and network of thin-walled blood vessels are also noted (H&E, ×40).

DISCUSSION

Most teratomas of infant and childhood arise in the sacrococcygeal region (2-4). Females are affected twice as often as males. Most sacrococcygeal teratomas diagnosed in infancy have low malignant potency and a good prognosis after excision. The frequency of malignancy in sacrococcygeal teratoma increases with age (5). The patient presented in this report was a 2-month-old baby girl with a grade II immature sacrococcygeal teratoma , who was born with the tumor.

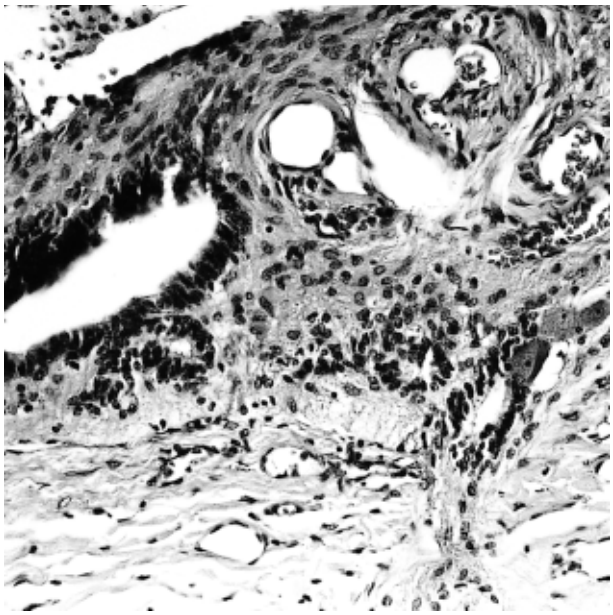

Fig. 2. Tubules of neuroepithelial type and ganglionic differentiation are apparent (H&E, ×200).

A wide variety of tumors, either benign or, more usually, malignant, may arise within teratomas. As many as 1 to 2 percent of mature teratomas harbor a cancer of adult. The most common of these is squamous cell carcinoma (6). Tumors of neuroepithelial origin may rarely arise within teratomas. The vast majority have been reported in the ovary and include neuroblastoma, glioblastoma, central neurocytoma, and primitive neuroectodermal tumors (7). In the sacrococcygeal region, we have found only one previous report of a teratoma with a component of a myxopapillary ependymoma (1).

The divergent neuroepithelial differentiation was studied in a transplantable mouse testicular teratoma (8). They included areas of mature primitive neuroepithelium with neuronal differentiation and mature neuroglial areas. The latter resembled astrocytoma, oligodendroglioma, and ependymoma. In our case, a large mass composed of oligodendrocytes was noted, which looked neoplastic transformation of clonal oligodendrocytes within the teratoma. Since the area composed of solely oligodendrocytes made definitive mass, it regarded as tumor rather than overgrowth.

Tumor-associated AFP may be elevated hundreds or thousands of times above the normal level in the serum. Elevated values generally portend a bad prognosis (9). In this case elevated level of serum AFP was found, which further increased after surgical excision of the tumor. Elevated AFP levels after surgery may indicate incomplete removal of the

tumor or the presence of metastasis or failure to chemotherapy.

Malignant sacrococcygeal teratomas are highly malignant tumors with an average postoperative survival of 18 months (10). In this case the patient died 6 months after the surgery due to intra-abdominal metastasis. We assume that the patient died of immature teratoma rather than oligodendroglioma.

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